Contents

Contents to Volume 63 (*Patients' Day*) xiv

Jules François (1907–1984) xix

Chris Ernst (1942–1986) xxi

Editorial xxiii

Introduction

E. Zrenner
Retinitis pigmentosa: a big challenge for scientists, physicians, and patients 3

H. Krastel
Clinical application of retinitis pigmentosa research: some recent examples 9

A. Laties
Overview of retinitis pigmentosa research 17

Classification and Clinical Aspects

J. François
Therapeutic approaches to retinal dystrophies caused by metabolic errors 23

R.W. Massof and D. Finkelstein
A two-stage hypothesis for the natural course of retinitis pigmentosa 29

S. Refsum, O. Stokke and O. H. Skjeldal
Refsum's disease (heredopathia atactica polyneuritiformis) — nosology, diagnosis, and treatment 59

W. Hammerstein
Fluoresceine angiographic findings in crystalline fundus dystrophy 69

W. Hammerstein, R.J. Seitz, K. Toyka and B. Schwartzkopff
Retinal dystrophy and mitochondrial changes 75

M. Ulbig, E. Zrenner and T. Schneider
Functional and morphological variations of fundus flavimaculatus 83

T. Schneider and E. Zrenner
Amplitude and implicit time changes in retinitis pigmentosa — an experimental approach 95

F. Genovesi-Ebert, G. Napolitano, M. Federghini, A. Oliva, G. Cavallacci and A.M. Wirth
Evaluation of hypothalamic dopaminergic activity in patients with retinitis pigmentosa 103

Short communication

J.G.H. Schmidt
Retinitis pigmentosa treatment: from disappointment to rational approaches 111

Tests of Visual Function for Differential Diagnoses

W. Ernst and A.T. Moore
Heterogeneity, anomalous adaptation and incomplete penetrance in autosomal dominant retinitis pigmentosa 115

W. Ernst, C.W. Tyler, G.C. Clover, B.A. Noble and D.J. Faulkner
Flicker studies of retinitis pigmentosa 121

H. Krastel and J.D. Moreland
Cut-off filters and the acuity luminance function in retinitis pigmentosa 129

B. Lorenz and E. Zrenner
Retinal dysfunction in hereditary myopia associated with choroidal degeneration. Is there a correlation to the degree of myopia? 137

E. Zrenner
Effects on cone function and cone interaction caused by drugs and congenital conditions that presumably increase cyclic AMP levels 155

C.M. Kemp
Fundus reflectometric investigation of retinitis pigmentosa 169

D. van Norren
Towards improved instrumentation for retinal densitometry 177

D.J. Faulkner, C.M. Kemp and S.G. Jacobsen
Fundus reflectometry and retinitis pigmentosa 181

K.A. Hellner and G. Walther
The early receptor potential (ERP) in retinitis pigmentosa 189

Short communications

B. Krogsaa, M. Warburg, M. Larsen and H. Lund-Andersen
Carriers of X-linked retinitis pigmentosa: a fluorophotometric determination of the blood–retinal barrier permeability 193

R. Kaufmann
The laser microprobe mass analyzer (LAMMA) as applied to ocular tissues. Cation- and trace element distributions in retinal cells 195

Molecular Genetics

A. *General*

T. Rosenberg and M. Schwartz
Genetic mapping of choroideremia 199

U. Friedrich, M. Warburg, P. Wieacker, T.F. Wienker and H.-H. Ropers
Linkage studies in retinitis pigmentosa 205

M. Geraghty, J. Farrar, J. Moloney, D.J. McConnell and P. Humphries
Linkage studies with L1.28 DNA probe on Irish families with X-linked retinitis pigmentosa 209

Short communication

A. Gal, T.F. Wienker and H.-H. Ropers
Early diagnosis of X-linked retinopathies by cloned DNA sequences 211

I.H. Pawlowitzki, A. Gall, D. Ahlert, H. Gerding, K. Rüther and F. Brunsmann
Genetic counselling in families with retinitis pigmentosa and choroideremia 215

B. *Workshop on molecular genetics*

H.-H. Ropers
Possibilities and limitations of recombinant DNA methods for the diagnosis of inherited disorders of the eye 217

G.J.B. van Ommen
Towards the detection and isolation of the retinitis pigmentosa gene 225

I.H. Pawlowitzki and F. Brunsmann
Choroideremia: the coordination of DNA banking by patient organization 239

M. Jay
The contribution of a genetic register to research 245

Biochemistry

A. *General*

M. Boulton, J. Marshall and J. Mellerio
Laboratory studies on human RPE cells isolated from donors with a variety of inherited retinal dystrophies 251

S. Klumpp, T. Pohl and J.E. Schultz
Cyclic GMP in the cilia of paramecium 265

S. Deshpande, R. Breuls, J.A. Parker, T. Borys and E.W. Abrahamson
Light scattering spectroscopic study of the proton ATPase activity in retinal dystrophic RCS rat 271

M. Boulton, W. Unger and J. Marshall
Functional and metabolic aspects of normal and dystrophic human retinal pigment epithelium in culture 281

G. Cremer-Bartels and K. Krause
Retinal pteridines 291

G.J. Chader
Biochemical studies of retinal degeneration in animal models and in the human 303

B. *Workshop on biochemistry of photoreceptors*

R. Paulsen
Introduction to the workshop on biochemistry of photoreceptors 323

R. Paulsen and K. Peters
Hereditary light-induced degeneration of a rhabdomeric photoreceptor 325

K.-W. Koch and U.B. Kaupp
Control of the light-regulated ion channel in vertebrate photoreceptors by cyclic GMP and L-*cis*-diltiazem 339

M. Wehner and H. Kühn
The cyclic GMP enzyme cascade of vision: site of light activation localized by enzymatic modifications of rhodopsin 345

S. Klumpp and J.E. Schultz
Regulation of cGMP-production in cilia from paramecium and tetrahymena by Ca^{2+}-ions: a model for vertebrate systems? 353

Morphology

A. *General*

J.M. Frederick, M.E. Rayborn, L.C. Moorhead and J.G. Hollyfield
Retinitis pigmentosa: correlative studies of histopathology and *in vitro* retinal metabolism in an autosomal dominant chorioretinal degeneration 367

El Sayed El-Hifnawi and W. Kühnel
The role of lysosomes in hereditary retinal dystrophy in RCS rats 381

M. S. Eckmiller
Photoreceptor outer segment renewal: special features in cones 397

H.H. Goebel and E. Dahme
Ultrastructural pathology of the retina in neuronal ceroid-lipofuscinosis of the Dalmatian dog 405

A. Lücke
Light damage and retinitis pigmentosa 411

El Sayed El-Hifnawi
Pathomorphology of the retina and its vasculature in hereditary retinal dystrophy in RCS rats 417

A.M. Meyer zum Gottesberge
Microanalytical investigations of the inner ear, uveal tract and retinal pigment epithelium melanin 435

B. *Workshop on animal models*

H.H. Goebel and E. El-Hifnawi
Introduction to the workshop on animal models 445

H.H. Goebel, N. Koppang, T. Bergsjø and E. Dahme
Canine retinal atrophy 449

Ch. Remé, Th. Hoppeler, A. Wirz-Justice and Ph. Hendrickson
Photoreceptor renewal systems: modulation by light and drugs 467

R. Lüllmann-Rauch and S. Lüpkes
Drug-induced degeneration of photoreceptor cells 471

Immunology

B. Detrick and J.J. Hooks
Immunologically related alterations in retinitis pigmentosa 479

O.F. Scheiffarth
Immunopathology of the posterior segment of the eye 489

B. Nölle and W. Müller-Ruchholtz
Anti-retinal antibodies in retinitis pigmentosa 497

M. Böhnke
Immunopathology of uveitis 507

A.A. Bialasiewicz
Drug management of intraocular inflammations. Some basic considerations 537

T. Lenarz, H. Krastel and W. Jaeger
Acquired sensorineural hearing loss and late manifestations of tapetoretinal disorders: clinical and immunological findings 543

Short communication

H. Krastel, T. Lenarz, W. Jaeger, E. Alexandridis, W. Tilgen and I. Anton-Lamprecht
Retinal, inner ear and skin dystrophic lesions of possibly autoimmune origin 553

Lipoprotein Metabolism

C.A. Converse, T. McLachlan, C.J. Packard and J. Shepherd
Lipid abnormalities in retinitis pigmentosa 557

I. Watanabe, M. Kato, H. Aonuma, A. Hashimoto, Y. Naito, A. Moriuchi and H. Okuyama
Effect of dietary alpha-linolenate/linoleate balance on the lipid composition and electroretinographic responses in rats 563

C.E. Jahn, O. Leiss, K.v. Bergmann and K. Schäfer
Serum lipoprotein concentrations in patients with retinitis pigmentosa 571

C.E. Jahn, K. Oette, A. Esser, K.v. Bergmann and O. Leiß
Increased prevalence of apolipoprotein E2 in retinitis pigmentosa 575

E.J. Schaefer
Diagnosis and management of ocular abnormalities in abetalipoproteinemia 579

Psychological Research

E.M. Glofke
Visual impairment by retinitis pigmentosa: coping with the crisis of becoming blind 585

Appendix 593
Discussions on immunotherapy in retinitis pigmentosa

Contributors' addresses 603

Index 611

Contents of Volume 63 (*Patients' Day*)

Foreword

Opening Addresses
The Federal Minister for Youth, Family and Health, Prof. Dr. R. Süssmuth
The Minister of Social Affairs for Hessen, A. Clauss
The Chairman of the Scientific Advisory Board of the German Retinitis Pigmentosa Society (DRPV), Prof. Dr. O.-E. Lund
The President of the International Retinitis Pigmentosa Association (IRPA), Mr. B. Berman
The President of the German Retinitis Pigmentosa Society, Dr. R. von Gizycki

Chapter 1
Researchers Help Patients — Patients Help Researchers
Towards the Cooperation between Researcher and Patients

F. Brunsmann and R.v. Gizycki
On the Current State of the Art and Development of RP Research from the Patient's Point of View

B. Berman
Challenging the Future

J. Mole
Emerging from the Tunnel

A. Christensen
A Total Registration of RP Patients and their Families in Denmark

G. Cousins
Establishing a Retinal Screening Centre, Patient Files, and Future Data Base

R. von Gizycki
The Promotion of Research by the German Retinitis Pigmentosa Society (DRPV)

Chapter 2
Technical Aids: Help Towards Autonomy

A. Blankenagel and U. Nechadi
Special Aids to Vision for Retinitis Pigmentosa Patients

W. Hammerstein
Indications for Vision Aids in Retinitis Pigmentosa

R. Bieker
Supplying Handicapped People with Technical Aids — Deficiencies and Ways of Tackling the Problems

R. Kefferpütz
A General Description of Closed-circuit Television and its Possibilities

D. Jäger
Advice on the Use of Residual Vision

E. Gerhard
Modern Aids: Computer Work Stations for the Partially Sighted and Blind

B. Steinke
The Supply of Technical Aids by Rehabilitation Organizations

W. Hammerstein
Advances and Developments in Technical Aids

Chapter 3
Therapy

H. Gussek
The Predicament of the RP Patient — a Dilemma

H. Krastel
Approaches to Therapy in Dystrophic Diseases of the Ocular Fundus

H. Hennekes
Methods for Monitoring Treatment in Retinitis Pigmentosa

U. Meyer
Problems in Drug Evaluation

E. El-Hifnawi
Animal Studies as Therapy Controls?

H.-R. Koch
Retinitis Pigmentosa and Cataracts

C. Jahn
Improved Diagnosis — a Key to Therapy

O.F. Scheiffarth
Immunotherapy in Retinitis Pigmentosa

C. Lane and M. Boulton
Retinal Pigment Epithelial Transplantation: Technique and Possible Application

Chapter 4
Genetics

I.H. Pawlowitzki and F. Brunsmann
Human Genetics — New Prospects?

M. Griffith
XLRP Breakthrough in Layman's Language

K. Rüther, F. Brunsmann, N. Ahlers, R. von Gizycki and I.H. Pawlowitzki
RP Patients and Molecular Genetic Research: a Study in Acceptability

Chapter 5
Living with RP
Family, Career, and Everyday Life

M. Mehtälä
An RP Patient's Description of Her Visual Disturbances

P. Cory
Maintaining Independence in Daily Living — Despite and With RP

B. Tasler
Living with RP — Patients' Mobility, Life Style, and Leisure Activities — Workshop Report

R. Rüberg
Our Lives Change Too — Partners and Parents of RP Patients

D. Wöhrle-Leberg
Teaching Programmes for Rehabilitating the Visually Handicapped, with Particular Emphasis on 'Orientation' and 'Mobility', as well as on 'Everyday Skills'

A. Larabi
Partially Sighted People in the Job Market — Opportunities and Limitations

M. Ersing
The Problems of the Visually Handicapped in Choosing a Career — Training and Retraining

P. Odenthal
The Institutional Framework Supporting Rehabilitation in the Federal Republik of Germany

Chapter 6
The RP Societies
Membership, Public Relations, Bookkeeping, Fundraising

H. Viitasaari
The New Members' Day — A New Activity of the Finnish RP Society

H.v. Velzen
How to Integrate Different Groups of Members into a Society

G. Kretschmer
The Main Aspects of the Use of Computers in the DRPV

B. Fasser
Cooperation between the RP and the Other Organizations for the Partially Sighted in Switzerland

E. Lehning-Fricke and W. Wehrhahn
The Current State of Public Relations Work in the DRPV

G.L. Duff
Motorcyclists Ride for Sight in Canada

Appendix

Constitution of International Retinitis Pigmentosa Association

Programme of the Congress

Scientific Advisory Board of the German Retinitis Pigmentosa Society (DRPV)

National RP Societies' Addresses

Contributors' Addresses

Index